

FACT SHEET

BOVINE SPONGIFORM ENCEPHALOPATHY (BSE) Q&A

What is Bovine Spongiform Encephalopathy?

Bovine Spongiform Encephalopathy (BSE), sometimes inappropriately referred to as "mad cow disease," is a chronic, degenerative, neurologic disorder of cattle. Affected animals display neurologic symptoms, such as changes in temperament (nervousness or aggression, incoordination, abnormal posture, loss of appetite, and decreased milk production. The incubation period (time of infection to time when signs start to show) is between two and eight years. The disease is always fatal, but time from onset of clinical signs to death is variable, and can be between two weeks and six months.¹

It is caused by an abnormal prion protein, which is transmissible. It is believed that when an animal is infected, the protein causes normal cellular proteins to change shape, and accumulate and damage the cells of the nervous system.²

There are two types of BSE: classical and atypical. The classical type (C-type) has been linked to variant Creutzfeldt-Jakob disease (vCJD) in humans, and is important because of this link.³ The atypical form of the disease occurs spontaneously at very low levels in cattle, is biologically different from C-type BSE, and therefore likely does not pose a risk to humans.

How is BSE spread?

The primary mode of transmission of C-type BSE is cattle eating feed contaminated with the infectious agent (prion protein). Animal feed can become contaminated if it contains the prion protein.

Does BSE exist in the United States?

According to the United States Department of Agriculture's (USDA) Animal and Plant Health Inspection Service (APHIS), "Five cases of BSE have been identified in the U.S. The first case in 2003 was confirmed as C-type BSE and the subsequent four cases were confirmed as atypical BSE forms.

The first case was detected in 2003 in Washington State in a 6-year-old dairy cow imported from Canada. The second, in 2005, was a 12-year-old beef cow in Texas. The third, in 2006, was a 10-year-old beef cow in Alabama. The fourth, in 2012, was a 10-year-old dairy cow in California. The fifth case, in 2017, was an 11-year-old beef cow in Alabama."⁴

Because the C-type has not been found in the United States since 2003, and both USDA and the Food and Drug Administration (FDA) have stringent animal import, food safety, and animal feed regulations and surveillance programs, the World Organization for Animal Health (OIE) upgraded the United States' BSE status to "negligible risk," which is the lowest risk any country can achieve.5

Why is BSE such a concern to the U.S. beef industry?

There are two main reasons that BSE is a concern in the U.S. beef industry. The first is that it is potentially a zoonotic disease, which means there is a chance that humans can be infected. With current food safety, animal feed, and animal import regulations, this chance is negligible. The U.S. government agencies continue to work with industry to institute surveillance and prevention strategies that ensure that U.S. beef is safe.⁶

The second reason it is a concern is that BSE is a trade-limiting disease. If a case of C-type BSE was found in the U.S., beef exports to other countries would come to a halt. This was evident in 2003, when the only C-type case of BSE was reported in the United States. The animal was destroyed, and government agencies increased regulation and surveillance activities. The OIE declared that the United States' BSE status was "negligible risk" in 2013, but trade restrictions continued for years, and were devastating to U.S. beef exports. Currently, global markets are again open to U.S. beef.



FACT SHEET

Does beef from BSE-infected animals make people sick?

No. No changes in beef consumption are necessary, and consumers should continue to enjoy beef and beef products as part of their diet. The BSE agent accumulates primarily in brain and spinal cord tissue in infected cows. In the United States, brain, spinal cord and other specified risk materials are banned for human consumption.

Does BSE pose a risk to humans?

Bovine Spongiform Encephalopathy is potentially a zoonotic disease, which means there is a chance that humans can be infected. The classical type has been linked to variant Creutzfeldt-Jakob disease (vCJD) in humans, and is important because of this link.⁸ With current food safety, animal feed, and animal import regulations, this chance is negligible.

Steps the U.S. has taken for prevention:

- USDA regulations prevent high-risk cattle and meat products from entering the U.S. from other countries which are at higher risk for BSE.9
- USDA food safety regulations only allow healthy cattle to enter the food supply. This includes a regulation that bans the slaughter of any cow that is non-ambulatory, or unable to rise and walk.¹⁰

- USDA food safety regulations also prohibit the use of high-risk tissue from cattle from entering the human or animal food supply. This high-risk tissue is called "specified risk material" or "SRM" and includes the brain, spinal cord, and other neurologic tissue. When cattle are slaughtered, this material is collected and destroyed.¹¹
- FDA does not allow most parts from cattle and some other animals to be used to make feed ingredients that are fed to other cattle.¹²
- Since 2006, USDA APHIS has implemented an ongoing surveillance system to monitor cattle and detect cases of BSE, whether they may be classical or atypical BSE. This surveillance program is designed to detect, with 95% confidence, one case of BSE per one million adult cattle.¹³

The U.S. government agencies continue to work with industry to institute surveillance and prevention strategies that ensure that U.S. beef is safe.

Helpful Links:

United States Department of Agriculture's Animal and Plant Health Inspection Service: https://www.aphis.usda.gov/aphis/ourfocus/animalhealth/nvap/NVAP-Reference-Guide/Control-and-Eradication/Bovine-Spongiform-Encephalopathy

World Organization for Animal Health (OIE): http://www.oie.int/index.php?
id=169&L=0&htmfile=chapitre bse.htm

References:

^{1,2,3,4,8,9}, ¹³ United States Department of Agriculture's (USDA) Animal and Plant Health Inspection Service (APHIS) BSE Website: https://www.aphis.usda.gov/aphis/ourfocus/animalhealth/nvap/NVAP-Reference-Guide/Control-and-Eradication/Bovine-Spongiform-Encephalopathy. Accessed June 19, 2019.

^{5,7} World Organization for Animal Health (OIE) List of BSE risk status. http://www.oie.int/animal-health-in-the-world/official-disease-status/bse/list-of-bse-risk-status/. Accessed June 19, 2019.

⁶,¹²United States Food and Drug Administration (FDA): All About BSE website. https://www.fda.gov/animal-veterinary/animal-health-literacy/all-about-bse-mad-cow-disease. Accessed June 19, 2019.

^{10, 11} USDA Food Safety and Inspection Services website: BSE Rules Being Strictly Enforced. <a href="https://www.fsis.usda.gov/wps/portal/fsis/topics/food-safety-education/get-answers/food-safety-fact-sheets/production-and-inspection/bse-rules-being-strictly-enforced/bse-rules-being-strictly-enforced. Accessed June 19, 2019.